Treatment of Syringomyelia in Patients with Arachnoiditis at the Craniocervical Junction

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■ OBJECTIVE: **Craniocervical junction arachnoiditis (CCJA) is an uncommon cause of syringomyelia. The pathophysiology of syrinx formation is uncertain, and the appropriate management unclear. A series of cases is reported to demonstrate variations in etiology, uniformity of functional cerebrospinal fluid obstruction at the foramen magnum, and results of surgical intervention.**

■ METHODS: **We retrospectively analyzed the clinical and radiologic features of a consecutive series of patients treated for syringomyelia related to CCJA.**

■ RESULTS: **Eight patients (5 male, 28–66 years old) were treated from 2000 to 2016. Magnetic resonance imaging demonstrated cervicothoracic syringomyelia in all cases, with the rostral extension of the syrinx suggesting communication with the fourth ventricle in all but one case. There was reduction of foramen magnum cerebrospinal fluid space in all cases, cerebellar ectopia in 5 cases, and fourth ventricular entrapment in 3 cases. Treatment consisted of posterior fossa decompression with either a GoreTex or pericranial patch graft. Six patients had a fourth-ventricle spinal subarachnoid shunt. Two patients had titanium mesh cranioplasty. The immediate postoperative period was associated with reduction in syrinx cavity size and improvement in neurologic symptoms in all cases. At follow-up 10–60 months postoperatively, 3 patients exhibited recurrence of the syrinx and underwent successful reoperation at the craniocervical junction. One patient with persistence of the inferior component of the syrinx was treated with a syrinx-spinal subarachnoid shunt.**

■ CONCLUSIONS: **Most syrinx cavities associated with CCJA communicate with the fourth ventricle. Posterior fossa decompression and fourth ventricle to spinal subarachnoid space shunting appears a reasonable treatment for this form of syringomyelia.**

INTRODUCTION

Syringomyelia is a disorder characterized by cystic cavities in the spinal cord containing fluid similar to cerebrospinal fluid (CSF) or extracellular fluid.[1-3] The exact pathophysiology of syringomyelia remains unclear, with elucidation complicated by the varied nature of associated conditions.[4] It occurs most commonly in association with Chiari malformation but also can be the result of spinal cord injury, intramedullary spinal tumor, spinal dysraphism, and spinal or craniocervical junction arachnoiditis (CCJA).[5,6] CCJA is a rare condition, occurring as a consequence of trauma, meningitis, subarachnoid hemorrhage, or previous posterior fossa surgery.[7,8] The treatment of syringomyelia associated with CCJA remains challenging, with recurrence rates exceeding 50%.[8] The authors present their experience in management of cervicothoracic syringomyelia associated with foramen magnum arachnoiditis.

METHODS

From January 2000 to September 2016, 125 patients underwent surgical management of syringomyelia by the senior author. All patients with CCJA from any cause, including posterior fossa surgery, were included. Patients with spinal arachnoiditis extending caudal to C1, or before surgery for Chiari malformation, were excluded. We retrospectively analyzed the clinical course,

Key words
- Arachnoiditis
- Craniocervical junction
- Fourth ventricle shunt
- Posterior fossa decompression
- Syringomyelia

Abbreviations and Acronyms
CCJA: Craniocervical junction arachnoiditis
CSF: Cerebrospinal fluid
MRI: Magnetic resonance imaging

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radiologic studies, and the surgical results of the cohort. This study was approved by the Macquarie University Human Research Ethics Committee.

RESULTS

Eight patients with cervical syringomyelia related to CCJA were identified, including 5 men and 3 women (mean age 43 years, range 28–66 years). All patients presented with upper-limb sensory and motor deficits (Table 1). One patient had cough-induced headache, and another presented with diplopia. The cause of arachnoiditis was trauma in 5 cases, viral meningitis in 1 patient, neonatal ventricular hemorrhage in 1 patient, and previous far lateral posterior fossa craniotomy for clipping of an unruptured posterior inferior cerebellar artery aneurysm in 1 patient. The cause of trauma was motor vehicle accident in 2 patients, birth injury in 2 patients, and sports injury in 1 patient.

Pretreatment magnetic resonance imaging (MRI) demonstrated cervicothoracic syringomyelia and attenuation of the cisterna magna and foramen magnum subarachnoid space in all cases. A diagnosis of CCJA was made preoperatively on the basis of dynamic MRI scans; however, the extent often is difficult to predict and was clarified intraoperatively. There were thick arachnoid bands intraoperatively in all cases. There was cerebellar ectopia in 5 cases and entrapment of the fourth ventricle in 3 cases. Cardiac-gated cine MRI sequences and phase contrast CSF flow studies were obtained in 4 patients, which demonstrated tethering of posterior fossa structures to the overlying dura, as well as limited movement of the cerebellar tonsils. These sequences showed no evidence of CSF flow in the dorsal subarachnoid space at the level of the cerebellar tonsils. Intraoperative ultrasound was used to assess the mobility of the posterior fossa contents and the architecture of the syrinx. In 1 patient, ultrasound revealed evidence of a direct communication from the fourth ventricle to the syrinx in the setting of fourth ventricular outflow obstruction (Figure 1). For 7 patients in all, the rostral extension of the syrinx was suggestive of communication with the fourth ventricle (Table 1).

Treatment in all patients consisted of posterior fossa decompression, division of adhesions, and expansile duraplasty. Duraplasty was performed with either a GoreTex Preclude MVP Dura (Gore & Associates, Flagstaff, Arizona, USA) substitute patch (3 patients) or autologous pericranial patch graft (5 patients). Six patients had a fourth ventricle-spinal subarachnoid shunt inserted. Titanium mesh plate cranioplasty was performed in 2 cases to allow hitching of the underlying dura substitute to the plate and expansion of the subarachnoid space (Table 2).

At mean follow-up 27 months postoperatively (range 10–60 months), clinical improvement had occurred in all cases. Specifically, preoperative symptoms of numbness, paresthesia, paresis, and spasticity consistently improved. Postoperative imaging demonstrated syrinx cavity size reduction in all cases. Three patients underwent reoperation due to syrinx recurrence (mean time to reoperation 32 months, range 24–39 months), entailing revision of the fourth ventricle to spinal subarachnoid space shunt in all cases, revision posterior fossa decompression in 2 cases, and revision duraplasty in 1 case. A separate syrinx to subarachnoid space shunt was performed in 1 case because of a persisting symptomatic syrinx in the thoracic cord 5 months after operation, despite collapse of the upper component. In all 4 cases, there was no further syrinx recurrence 15–52 months after reoperation.

ILLUSTRATIVE CASES

Four cases from the series exemplify the 3 major causes, the shared radiologic features, as well as our treatment strategies in syringomyelia associated with CCJA.

Patient 1: Posttraumatic CCJA Due to Motor Vehicle Accident

This 35-year-old man presented with worsening cough-induced headaches, limb paresthesia, and global weakness and hyperreflexia of the limbs 13 years after an occiput-C1 fusion after a motor vehicle accident. MRI revealed a large cervical syrinx and descent of the cerebellar tonsils to C1 (Figure 2).

The patient underwent posterior fossa decompression, with suboccipital craniectomy. The posterior arch of C1 already had been removed. There was dense epidural scarring and calcification, which was removed while the fusion was left intact. The dura was opened in the midline, and extensive arachnoid adhesions

Table 1. Presenting Symptoms and Pathogenesis

Patient	Age, years	Sex	Symptoms	Pathogenesis of CCJA	Syrinx Location
1	35	Male	Headaches, sensory disturbance of limbs	MVA trauma	C1–C7
2	40	Female	Upper-limb sensory disturbances and weakness	Meningitis	C1–T5
3	66	Male	Limb sensory disturbances, weakness, diplopia, and respiratory depression	Postsurgical	C1–T1
4	36	Male	Right-sided facial pain, right upper limb sensory disturbance without weakness	Birth trauma	C1–C7
5	32	Female	Upper-limb sensory disturbances and weakness	MVA trauma	C1–T9
6	54	Male	Headache, upper-limb sensory disturbances and weakness	Sporting trauma	C3–T9
7	28	Male	Right upper limb sensory disturbances without weakness	Neonatal ventricular hemorrhage	C1–T4
8	55	Female	Left arm and shoulder pain and paresthesia	Birth trauma	C1–C4

CCJA, craniocervical junction arachnoiditis; MVA, motor vehicle accident.